Case Report

# Unusual achalasic sigmoid esophagus

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## Abstract

Achalasia cardia is a rare primary esophageal motility disorder with an incidence of about 1 in 100,000 individuals. If left untreated or inappropriately treated, the disease may progress to end-stage achalasia characterized by megaesophagus or sigmoid esophagus. The presentation as severe malnutrition, anemia, hypoalbuminemia, and asymptomatic tracheal compression has not been described in the setting of the sigmoid esophagus, in absence of underlying malignancy. Here we report an interesting case of a young male, who presented with the above features and managed excellently by open Heller's myotomy under epidural anesthesia due to the poor performance status.

**Keywords:** Achalasia cardia; Heller's myotomy; Malnutrition; Sigmoid esophagus.

## Introduction

Achalasia cardia is a rare primary esophageal motility disorder with an incidence of about 1 in 100,000 individuals.[1] It is caused by selective degeneration of inhibitory neurons of the esophageal myenteric plexus, leading to a lack of peristalsis and the non-relaxing lower esophageal sphincter (LES). This eventually affects the emptying of food from the esophagus into the stomach and causes dilatation and tortuosity of the esophageal body.[2] If the patient is left untreated or inadequately treated, the disease may progress to end-stage achalasia characterized by megaesophagus and/or sigmoid shaped esophagus.[3] End-stage achalasia is even a rarer entity, comprising only 4% of all achalasia.[3]

Although unintentional weight loss is common, severe malnutrition is very rare unless there is an underlying malignancy.[1,4] Here we report an unusual case of a young patient with sigmoid esophagus who presented with a short duration history of dysphagia with three major complications of severe malnutrition, anemia and tracheal compression in the absence of underlying esophageal malignancy.

## Case Report

A 35-year-old male presented with complaints of dysphagia to both solid and liquid for 4 months. He also gave a history of vomiting and regurgitation of food following each meal and a significant weight loss of around 15 kg. The patient denied any history of chest pain, shortness of breath and luminal bleeding. On examination, the patient was cachectic, dehydrated and pale with a body mass index (BMI) of 12.7 kg/m$^2$. The pulse rate was 110 beats/min, blood pressure of 90/60 mm Hg with normal body temperature. The chest auscultation was bilaterally clear, the abdomen was scaphoid with no other significant findings. Blood investigation showed a low hemoglobin (5.5 gm/dl; range: 12-14 ), leucocytosis (21,800 cells/mm$^3$ ; range: 4000-11,000), deranged renal function test and serum biochemistry ( urea- 64 mg/dl; range: 10-40, creatinine- 0.7 mg/dl; range: 0.5-1.4, Sodium- 140 meq/l; range: 135-145, Potassium- 2.9 meq/l; range:3.5-4.5), and hypoalbuminemia (2.3 gm/dl; range: 3.5-5.0). Chest x-ray showed mediastinal enlargement with an air-fluid level. Barium swallow revealed a smooth narrowing in the distal esophagus with gross proximal dilatation. Similarly, endoscopy revealed dilated esophagus containing food residue with multiple ulcers. The mucosa of the distal esophagus and cardia of the stomach were normal. However, in view of short duration history and severe malnutrition with underlying anemia, malignancy (pseudoachalasia) could not be ruled out and we requested contrast computed tomography (CT) of the chest and abdomen, which showed a hugely dilated and tortuous esophagus (11 cm) containing

food residue, compressing the trachea, and excluding underlying malignancy (occult adenocarcinoma of cardia or metastatic lymph nodes at gastroesophageal junction) (Figure 1). Based on this, the diagnosis of achalasia cardia with sigmoid esophagus with an Eckardt score of 10 was made and planned for surgical intervention.

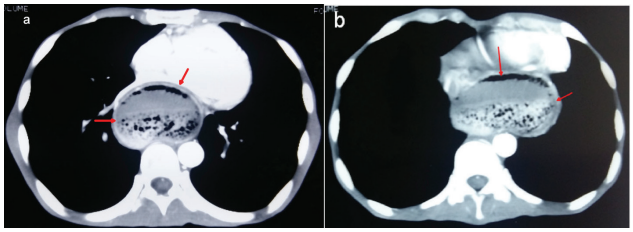


**Figure 1. Contrast CT chest showing hugely dilated esophagus (11cm size), containing food residue *(arrow)* coursing from the right *(a)* to the left side *(b)* of the chest.**

The patient was initially resuscitated with intravenous fluids and an empirical intravenous antibiotic. A nasogastric tube was inserted for decompression and lavage of the esophagus. He received three units of packed red blood cells. Central venous catheter was placed and parenteral nutrition was started and continued for the next 10 days which led to an overall improvement in his general condition. In spite of nutritional supplementation and stabilization, the patient was not fit enough to tolerate general anesthesia, hence in collaboration with anesthetics, we opted for open Heller's cardiomyotomy with Dor's fundoplication under thoracic epidural anesthesia. The epidural block was achieved by 2 ml aliquots of 0.5% ropivacaine (total dose 10 ml) at $T_{10/11}$ interspinous space. Operative findings showed a dilated esophageal segment (11cm) with a 2 cm long thickening in the distal esophagus. No growth or foreign body could be identified. The operative procedure was uneventful with an operating time of 45 minutes and the length of myotomy was 8.0 cm (Figure 2).

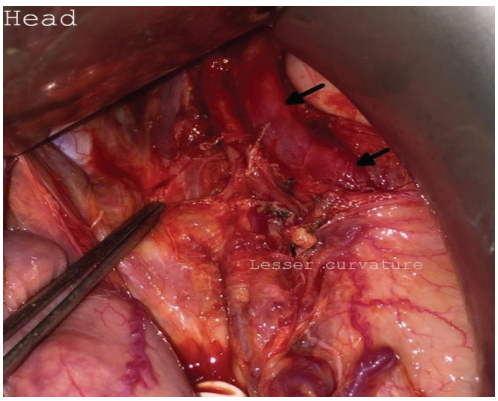


**Figure 2. Intraoperative photograph showing bulging mucosa *(arrow)* of the esophagus following adequate myotomy.**

A liquid diet was allowed at postoperative day 1 and was gradually changed to a regular diet at postoperative day seven. The postoperative Eckardt score dropped to 2, suggesting an improvement. At two years of follow up, the patient has gained weight (12 kg) and enjoying normal swallowing of all kinds of food with a good quality of life. His follow-up chest x-ray shows residual esophagus dilatation, without any dysphagia/regurgitation (Figure 3).

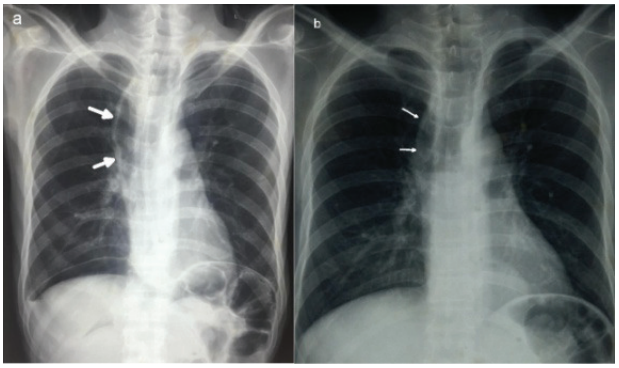


**Figure 3. Follow-up chest radiograph, immediate *(a)* and at three months *(b)* visit showing residual esophageal dilatation *(arrow).***

## Discussion

The most common presentation of achalasia cardia is dysphagia to both solids and liquids followed by heartburn, regurgitation and chest pain.[1] As the esophagus gets progressively dilated and turns to end-stage disease, nocturnal cough, regurgitation/aspiration and weight loss predominates.[5] They may frequently present with recurrent pneumonia and even (rarely) with tracheal compression causing respiratory distress due to the compressive effect of megaesophagus, which was however not seen in the present case.[6] Due to the retention of esophagitis and esophageal ulcers, they may bleed causing anemia. Severe malnutrition, weight loss, and anemia are unlikely without an underlying malignancy, as most of the patients are well-preserved in spite of dysphagia due to stereotyped movements they adapt with meals.[7] Our case was rare and unique in the sense that, it presented with a rare combination of severe malnutrition, anemia requiring a blood transfusion, asymptomatic tracheal compression, hypoalbuminemia and feature of sepsis due to probable microaspiration of retained esophageal food.

The treatment of patients with achalasia cardia and megaesophagus/sigmoid esophagus remains controversial. Traditionally, the management of this so-called "end-stage achalasia" has been esophagectomy. This was based on the perception that the esophageal body peristalsis

would not be able to empty effectively even after an adequate myotomy. Moreover, there is usually significant periesophageal inflammation, esophagitis, and ulceration due to the longstanding retention of food making myotomy a difficult procedure.[8,9]

Nowadays, this notion has been challenged, and recent studies have demonstrated symptoms improvement in more than 90% of patients treated with laparoscopic Heller's myotomy (LHM) with an endotracheal intubation.[8] Esophagectomy, should be reserved as a last resort in patients in whom all other modalities have failed or require resection because of concomitant esophageal malignancy.[3,9] In a patient unfit for general anesthesia (poor performance status), with a tracheal compression due to the sigmoid esophagus as seen in our case, an open Heller's myotomy under thoracic epidural anesthesia is also an alternative.

**Conflicts of interest**

The authors of this case report have no financial or ethical conflicts to disclose.

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